

Case Report

Sudden death due to acute pulmonary embolism from asymptomatic right atrial myxoma

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Abstract

A 21-year-old man, in whom abnormal nodules had been detected in bilateral lung fields with no clinical symptoms for two years, was admitted to the hospital with sudden cardiopulmonary arrest. Acute pulmonary embolism with a large embolus was diagnosed, but the patient died soon after admission. When the examination was compared with X-ray taken 4 days earlier by chance, a large tumor was now detected in the right heart and was suspected to be the cause of this complication after his death. Necropsy found a $3.8 \times 3.5 \times 1.0$ cm myxoid tumor arising in the right atrium and a large fragment of this type of tumor was at the pulmonary trunk. Many old myxoma fragments were noted in the bilateral peripheral branch of the pulmonary artery. It was concluded that the abnormal nodules were old pulmonary fragments and the cause of death was pulmonary embolism of a large fragment originated from the atrial myxoma. An asymptomatic right atrial myxoma is extremely rare but nevertheless possible to unexpected death like this case.

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1. Introduction

Myxomas account for 50% of primary cardiac tumors and are usually found in the left atrium. A right atrial myxoma is a rare finding.¹ Generally, the tumor has been found with symptom related to pulmonary embolism, cardiac obstruction, and constitutional disturbance, and rarely with no clinical symptom.^{2–4} We report here on an atypical case of a right atrial myxoma which was discovered as a case of sudden onset of pulmonary embolism with no prior clinical symptoms.

2. Case report

A 21-year-old man was admitted to the hospital with sudden cardiopulmonary arrest. Multiple nodules in bilat-

eral lung fields had been detected two years earlier. However, because the patient had no subjective symptoms, he had not been subjected to a thorough examination yet. At 4 days before admission, he had made a medical examination following annual check up in a hospital by chance. His chest X-ray was taken (Fig. 1, Former), and a doctor recommended him a further examination for the multiple nodules in the lungs without noting the shadow on his heart. He reserved it at 7 days after and spent his daily life. Chest X-ray taken after crisis revealed a large fragment in a pulmonary trunk causing a pulmonary embolism (Fig. 1, Later). Despite resuscitation including the thoracotomy to exclude fragments from the pulmonary artery trunk, he expired soon after admission. When compared with his earlier chest X-ray, a large tumor in the right heart was now discovered along with multiple pulmonary nodules (shown as large arrow in Fig. 1, Former) and was suspected to be the cause of this complication.

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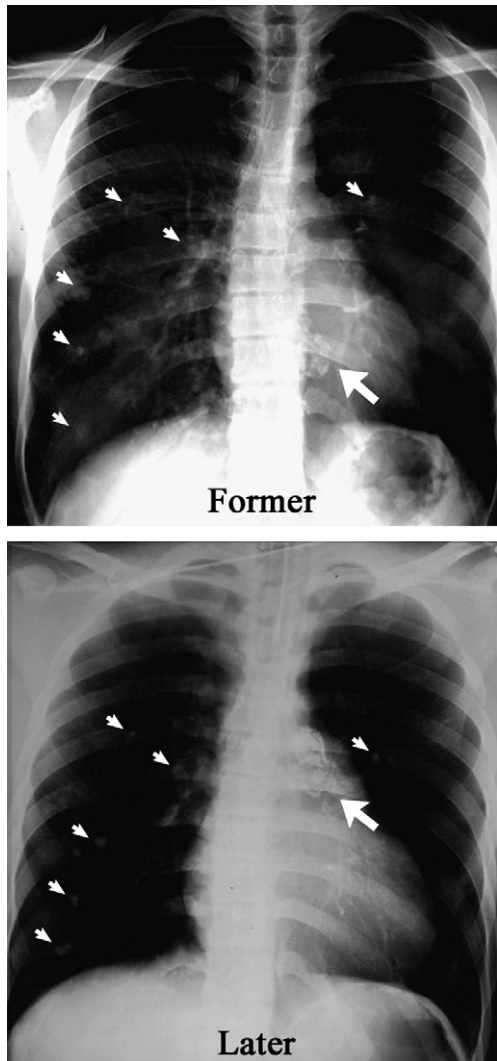


Fig. 1. Chest X-rays. *Former*: A large tumor in the right heart (large arrow) appears along with several pulmonary nodules in the bilateral lung field, especially in the right lung field (small arrows) taken at 4 days before admission. *Later*: A large fragment of the tumor moves to pulmonary trunk (large arrow) causing pulmonary embolism taken right after the crisis.

At necropsy practiced on the next day, a large papillary tumor ($3.8 \times 3.5 \times 1.0$ cm) with gelatinous consistency was found in the right atrium (Fig. 2A). The tumor arose from the margin of the fossa ovalis and was very friable. Hypertrophy or dilatation of the right ventricle was not obvious. Histologically the tumor consisted of an acid-mucopolysaccharide-rich stroma, polygonal cells with scant eosinophilic cytoplasm, fibrocytes, smooth muscle, and lymphocyte. Also, frequent calcifications were observed in the matrix (Fig. 2B). These findings were typical of myxoma. The pulmonary trunk was filled with a fragment in which same property was observed as the right atrial myxoma with naked eye associated with histopathological correspondence. The lung felt abnormally firm and bled widely when sectioned. Some pulmonary arteries, from the main lobar branches to the smallest branches, were occluded by a firm

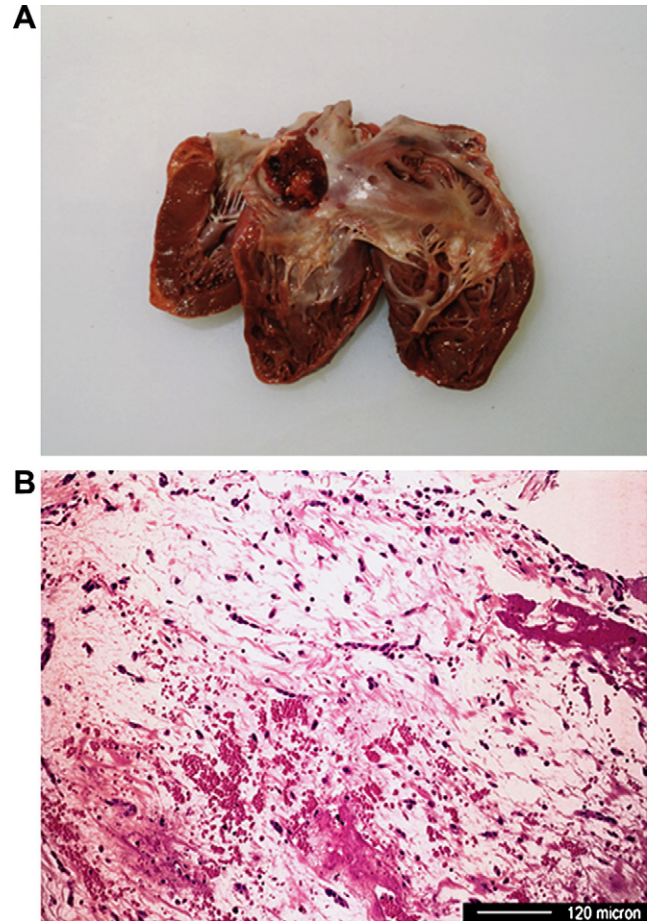


Fig. 2. Papillary tumor of right interatrial septum at the margin of the fossa ovalis (A). Histologically, the tumor consists of an acid-mucopolysaccharide-rich stroma with hemorrhage. Polygonal cells with scant eosinophilic cytoplasm and other cellular elements such as fibrocytes, smooth muscle, lymphocytes and calcifications can be observed in the matrix (B). Hematoxylin–Eosin stain.

and jelly-like substance corresponding with the abnormal nodules pointed out in the earlier chest X-ray (Fig. 1, *Former*). These nodules were identified as myxoma covered with fibrous capsule of various thicknesses, with the findings indicating the repeated embolism since before.

3. Discussion

This is a report on a large right atrial myxoma with multiple pulmonary emboli with no physiological symptoms. Cardiac myxomas are thought to arise from embryonic rests, which may differentiate into several cell types.⁵ Histologically, they consist of a myxoid matrix composed of an acid-mucopolysaccharide-rich stroma. Polygonal cells arranged singly or in small clusters are scattered throughout the matrix. Fibrocytes, smooth muscle, lymphocytes and plasmacytes can be seen occasionally in the matrix.^{5,6} In this case, the structures were consistent with above-mentioned myxoma, and calcifications were frequently observed (Fig. 2B). Reynen et al. reported that calcification

occurred in 10% of cardiac myxomas.⁷ Calcifications might be derived from postnecrotic degeneration in the tumor as most of calcinosis were so, which followed the tumor old at least several months or more, while detailed mechanisms were not reported in the paper.⁷

Most of the myxomas arise from the interatrial septum and are polypoid with a smooth or gently lobulated surface. Less commonly they are villous or papillary with very fine villi which are gelatinous and fragile, easily breaking off into pieces.^{6,7} The myxoma in this report arose from the interatrial septum at the margin of the fossa ovalis and was papillary, gelatinous and finely friable (Fig. 2A). At necropsy, a large fragment of the myxoma was found in the pulmonary trunk and some fragments of myxoma covered with fibrous capsule of various thicknesses were found in the bilateral peripheral branch of pulmonary arteries corresponding to the abnormal nodules pointed out previously. Due to these findings, it was concluded that the nodules were derived from the fragments of the fragile tumor which had been recurrently embolized in the peripheral vessels since before. Heath et al. demonstrated an embolized fragment of myxoma invading the adventitia,⁸ and believed the myxoma tissue proliferated and invaded the occluded vessels. It might be concerned with the present case, but we were unable to confirm these sequential events in the fragile tumors.

The clinical symptoms of right atrial myxoma include the triad of pulmonary embolism, intracardiac obstruction, and constitutional disturbance such as fatigue, fever and weight loss.^{2–4} A right atrial myxoma very rarely causes pulmonary embolism by fragments. In most clinical reports, a right atrial myxoma gives rise to signs of cardiac obstruction with dyspnea, syncope and general edema.^{1,9–11} These symptoms are related in part to the size of the tumor. There are no symptoms with a small tumor, but large tumors can cause an obstruction of the tricuspid valve and right atrium, resulting in right sided heart failure. In our cases, there were no findings at necropsy of right sided heart failure such as right ventricular hypertrophy, splenomegaly and leg edema. The patient had no clinical symptoms in spite of a sufficiently large tumor in the right heart and old fragments of the tumor (Fig. 1, Former). Several investigators demonstrated that the clinical features of

myxoma were affected by mobility in addition to size.^{5,7} Due to the calcifications noted, the tumor in this report was slightly hard and consequently had a mobility defect. This may have contributed to the lack of clinical symptoms until the tumor moved to fully embolized the trunk of pulmonary artery at the first cardiopulmonary attack.

In conclusion, a case of right atrial myxoma is described, which was asymptomatic in spite of the presence of a large intracardiac tumor and multiple old fragments in the pulmonary artery tree. In such a rare case, a right atrial myxoma may nevertheless account for sudden unexpected death.

Conflict of interest statement

None of the authors have any conflicts of interest associated with our report.

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